Fenestrated Anterior Cerebral Artery with Associated Arterial Anomalies

Case Reports and Literature Review

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Summary

Fenestration of the A2 segment is extremely rare. Cerebrovascular fenestration may be associated with an increased incidence of cerebral aneurysm and other vascular anomalies. Two case reports are presented which identify a fenestration of the A2 segment and other normal variations of the intra-cerebral circulation.

A review of the literature has been undertaken to determine the prevalence and embryology of anterior cerebral artery fenestrations, their clinical significance and the association with aneurysm formation and other intracranial vascular anomalies.

Case Reports

Case 1: A fenestrated A2 segment was demonstrated on both CT angiogram and angiography in a 62-year-old woman, who initially presented after a syncopal episode while driving resulting in a high speed motor vehicle accident. A small subarachnoid haemorrhage was noted on a non contrast CT scan of the brain with blood within the sulci in the right frontoparietal region. At the time of presentation, this was deemed to be a traumatic subarachnoid haemorrhage and was not further investigated during the initial admission. The patient was reviewed by a neurologist three months later who ordered a CT angiogram.

Multi-detector CT (MDCT) angiogram and subsequent formal digital subtraction angiography demonstrated a 3 mm aneurysm of the anterior communicating artery (AComA). In addition to the fenestrated A2 segment, several vascular developmental anomalies were also demonstrated including a fenestrated AComA, a hypoplastic left A1 segment and a right hypoglossal artery (Figures 1-3). The aneurysm of the AComA was treated by balloon assisted coil embolization one month later due to the possibility that it was the source of the subarachnoid haemorrhage. The patient was discharged without neurological compromise.

Case 2: A 42-year-old woman presented to the emergency department with a sudden onset of a severe headache. The patient's father had died aged 34 of an atraumatic intra-cerebral haemorrhage. The cause for this haemorrhage had not been identified. MDCT angiography of the neck and Circle of Willis identified a fenestrated right A2 segment. Other normal variations that were identified included the left A2 segment arising from the proximal A1 segment and a hypoplastic right A1 (Figure 4). No intracerebral aneurysm was identified. The patient did not undergo formal angiography.

Discussion

A fenestration has previously been described as the division of the arterial lumen, with resulting separate channels, each with its own en-

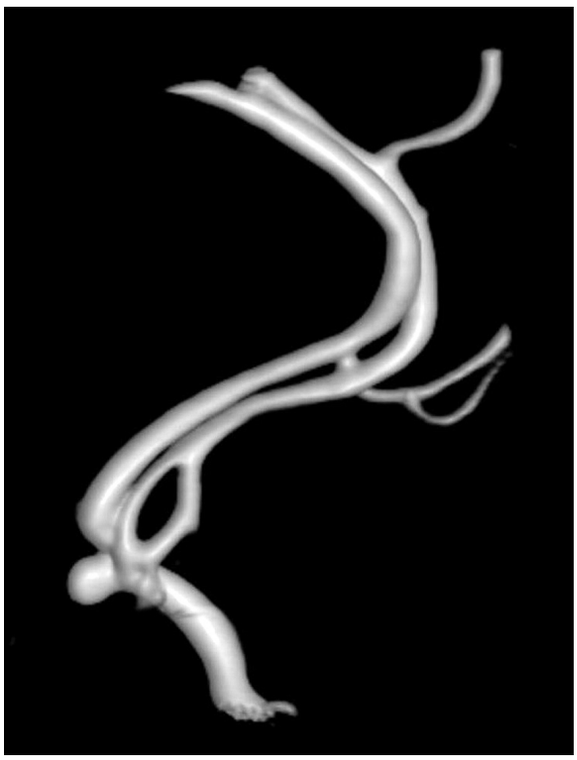

Figure 1 3D reconstruction of the right ICA angiogram (case 1) demonstrating the A2 segment fenestration and an aneurysm arising from the anterior communicating artery.

dothelial and muscularis layer, however, these separate arterial channels may or may not share the adventitia[1]. Fenestrations are more common in the vertebrobasilar arteries[2].

Fenestrations of the anterior cerebral artery (ACA) are predominantly observed within the distal A1 segment[3] or the anterior communicating artery. The incidence of fenestration of the A1 segment is between 0-4%[4] on anatomical studies and 0.058% on angiographic studies[2]. The reported incidence of fenestration of the AComA is 5.3% on angiography[5] and 7.5-40% in autopsies[6].

Fenestration of the A2 segment has been described anatomically by Vucetic[7] during dissection of 200 fetuses at 20-40 weeks gestation. Four cases were demonstrated in this study. Uchino et Al[8] provided a brief description of one case of fenestrated A2 on MRI angiography. Unfortunately, no imaging of this finding was provided and little detail is offered by the authors to allow comparison with our reported cases. They describe the fenestration as small and slit-like. The subject in case 1 of this paper had an ovoid shaped fenestration. A single case of a slit-like fenestration in an azygous A2 was also described by Uchino el Al[8].

The aetiology of fenestrations of the A1 and A2 segments is not well understood[3,9]. In embryos of 4-5.7 mm (28-30 days), the cranial and caudal divisions of the internal carotid artery (ICA) are formed[10]. The cranial division of the ICA constitutes the primitive olfactory artery, which in turn, gives off the anterior choroidal and middle cerebral arteries[11]. In embryos of 11.5-18 mm (41-48 days) the primitive olfactory artery has two branches, the main (original) vessel to the nasal fossa, and a secondary, more medial vessel which represents the future continuation of the ACA[11]. At the end of this stage, the more medial artery is joined by its counterpart on the opposite side by a plexiform anastomosis. The networking vessels in this anastomosis gradually coalesce to form a single AComA. Padget[12] demonstrated fenestrations in this region in a 14-mm embryo, with persistence of the fenestrations in 18- and 24-mm embryos. The fenestrations, however, were not present in the 43-mm embryo.

The primitive olfactory artery also participates in the plexiform anastomosis but gradually disappears. It is postulated that the fenestrations in this region are remnants of the plexiform anastomosis, especially between the primitive olfactory artery and the ACA[12,13].

Unfortunately, this hypothesis may possibly only be applied to fenestrations of the A1 segment and AComA. Consideration of other normal variants of the A2 segment may also assist in determining an embryological cause for fenestrations of this artery. These variants include an azygos A2 and trifurcation of the A2 segment.

An azygos ACA is a more common normal variant of the A2 segment which represents a single trunk that supplies both hemispheres. The incidence of azygos ACA ranges from 0 to 5%[14]. Padget described a vessel sited at the midline at the 24-mm stage that she called the median artery of the corpus callosum which arises from the AComA, courses parallel to, and behind the normal pericallosal artery and supplies the corpus callosum, septal nuclei, septum pellucidum, rostral portions of the fornix, and a portion of the frontal lobes[10,12]. This vessel normally involutes as the ACA segments distal to the AComA develop. The formation of an azygos A2 segment is thought to result from

abnormal fusion of the paired A2 from the medial branch of the primitive olfactory artery at the 16-mm stage of embryonic development, or because of persistence of the median artery of the corpus callosum at the 20- to 24-mm stage and regression or lack of development of the ACAs [12,15,16]. The definitive aetiology of an azygos A2, however, remains uncertain [10].

Trifurcation of the A2 segment is another common normal variant which is defined as three A2 segments. The reported incidence of a trifurcation of the A2 segments arising from the AComA varies from 2 to 13% [17]. This normal variant most likely represents two normal A2 segments and an additional vessel which is a persisting embryonic median artery of the corpus callosum.

It may be postulated that anastamosis of the proximal portion of the median artery of the corpus callosum and regression of the remainder of the artery with the proximal A2 segment may result in an A2 fenestration.

Krigg et Al (2007) recently proposed a revised nomenclature which defines fenestrations, segmentally unfused arteries and duplications. A fenestration is defined as a single artery with two luminal channels and may be due to a nerve or other anatomical structures "piercing" the artery. These are typically encountered in the vertebral artery or the ICAs in the neck [18].

Segmentally unfused arteries constitute a lack of fusion of embryologically paired vessels. Duplications can occur where the "double lumen" is due to two embryologically different vessels that fuse during development and an additional vessel persists. This is distinct from a fenestration where the two lumina correspond to a single artery [18].

Duplications are reported to occur in the ICA (segmental agenesis of the first ICA segment with reconstitution of the distal ICA via different arteries from the ascending pharyngeal artery system), the ACAs with duplicated vessels and persistent infraorbital origin of the ACAs or at the vertebrobasilar junction [18].

Using this nomenclature, Krings et Al reported a case of a segmental unfused basilar artery which would previously have been described as a fenestration. Embryologically, this anomaly fits the definition as there is failure of fusion of the longitudinal arteries which form the basilar artery [12]. However, due to the uncertainty of the embryological aetiology of the same anomaly within the anterior cerebral arteries an exact label is difficult. If, for example, A2 fenestration is due to remnants of the plexiform anastomosis, especially between the primitive olfactory artery and the anterior cerebral artery then this anomaly would more appropriately be described as an unfused A2 segment. However, if this anomaly arises due to

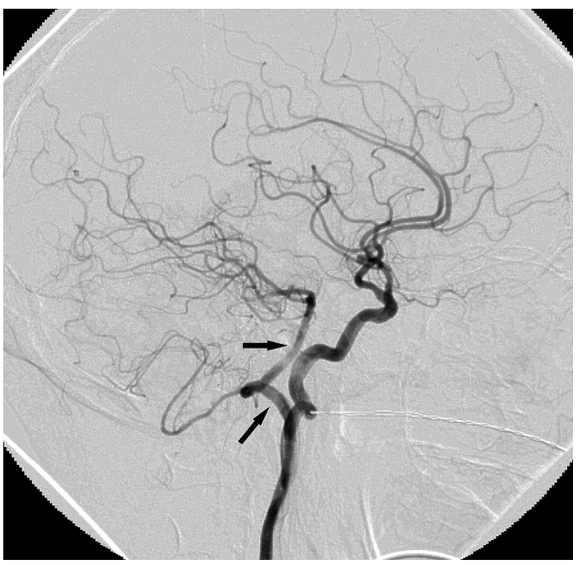

Figure 2 Frontal projection of right ICA angiogram (case 1) demonstrates the persistent hypoglossal artery (black arrows).

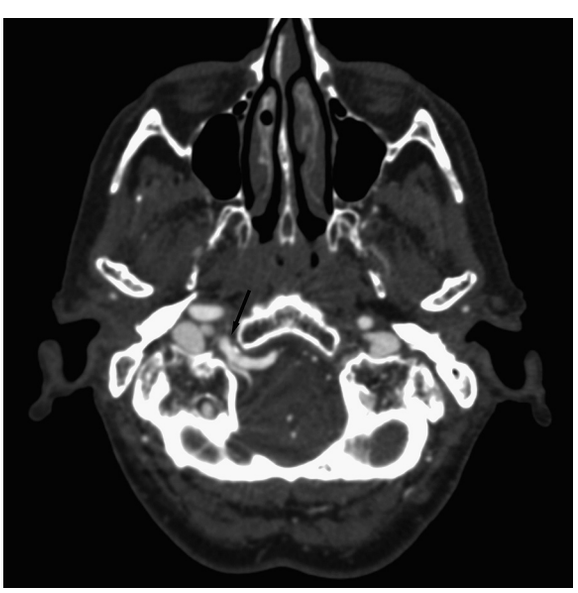

Figure 3 Axial image obtained from MDCT angiogram (case 1) shows the right hypoglossal artery entering the intra-cranial cavity via the hypoglossal canal (black arrow).

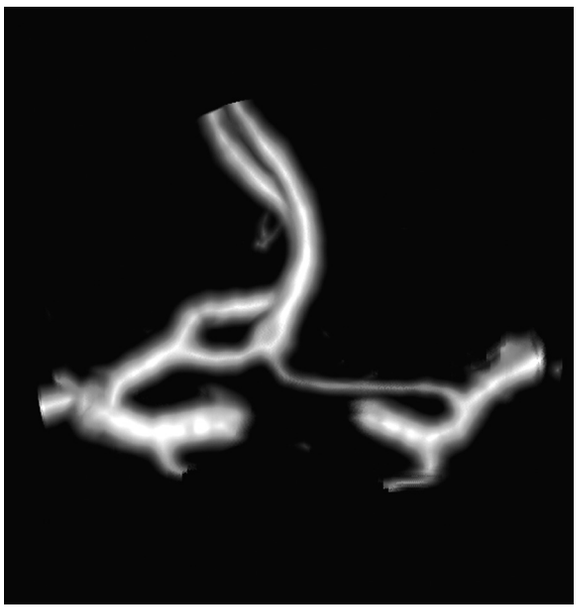

Figure 4 PA projection of 3D image from MDCT angiogram (case 2) shows a fenestrated right A2 segment, hypoplastic right A1 segment and the proximal left A2 arising proximally from the A1 segment.

an attachment to a median artery of the corpus callosum, then the term duplication is best applied. The nomenclature proposed by Krings et Al therefore may not be as easily applied to a "fenestration" of the A2 segment.

Case one of this report demonstrated a 3 mm aneurysm associated with a fenestration of the AComA. One third of all cerebral aneurysms arise in the region of the AComA complex [11]. Most fenestrations, however, occur in the posterior circulation and are not associated with aneurysm formation. Some authors therefore postulate that intracranial fenestrations do not have a higher risk of aneurysm formation [2,11]. Other authors report that ACA fenestration is frequently associated with an aneurysm at the proximal end of the fenestrated segment and therefore hypothesize there is a definite link [22].

The lateral walls of a fenestrated artery have a normal intrinsic architecture. However, the media layer is absent at the base of the medial wall and in addition, the subendothelium is thin and there is discontinuity of the elastin layer [23,24]. Cerebral artery bifurcations have been demonstrated to have a similar structural anatomy [24,25]. It has been postulated that sites of fenestrations may represent an incomplete maturation process of the arterial wall and contain "weaker", less mature endothelial cells, which may later develop arterial aneurysms [26]. A further hypothesis for the formation of aneurysms at sites of fenestration is the defect of the tunica media at the proximal end of the duplication may create turbulent flow at the site of bifurcation [1]. Uchino et Al [8] reported that 14% of AComA aneurysms are associated with a hypoplastic A1. They postulated that AComA aneurysms frequently occur in patients with an asymmetric A1 segment due to haemodynamic stress [8].

Multiple further aetiologies have been suggested for the development of intracranial aneurysms. These include haemodynamic stresses, genetic and environmental factors. Genetic predisposition has been linked to both a locus on chromosome 17 [19], a linkage site on chromosome 3 [19] and polymorphisms within the kallikrein gene cluster [20]. Certain disorders may also predispose an individual to develop intracranial aneurysms. These include autosomal dominant polycystic kidney disease (chromosome 16) and fibromuscular dysplasia [21].

Case one of this report also demonstrated a persistent hypoglossal artery. The persistent hypoglossal artery is the second most common carotid-basilar anastamosis [6]. The incidence of this anomaly is estimated as 0.027-0.26% [1]. A persistent hypoglossal artery may be associated with an increased incidence of aneurysm. Persistent primitive anastomoses are postulated to be associated with a higher risk of cerebral aneurysm formation, both involving the persistent embryological vessel as well as occurring elsewhere in the cerebral vasculature. It may be hypothesized that a cluster of vascular abnormalities and aneurysms may be secondary to halting of the normal cerebrovascular development at a primitive stage indicating an underlying genetic vascular abnormality. The aetiology of intracranial aneurysm formation may therefore be more complex than simple flow dynamics alone.

Conclusions

This paper presents the radiological findings of two cases of a rare fenestration of the A2 segment of the anterior cerebral artery. On review of the literature, the aetiology of fenestrations of the anterior cerebral artery remain unclear and there is no consensus whether fenestrations of intracranial arteries predispose individuals to aneurysm formation.

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